Polyarteritis nodosa presenting with frank hematuria

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Abstract

Polyarteritis nodosa (PAN) is an uncommon systemic vasculitis characterized by necrotizing inflammation of small- or medium-sized arteries. The disease normally presents with non-specific symptoms. Urological symptoms at presentation are extremely rare. We report a 65-year-old man who was diagnosed with a polyarteritis nodosa having presented atypically with left testicular pain and swelling, and an intratesticular lesion. He developed painless visible hematuria while under investigation. No gross arterio-venous fistula was seen to suggest a false aneurysm. Subsequently, laboratory studies showed positive anti-neutrophil cytoplasmic antibody levels and a raised erythrocyte sedimentation rate. This was an unusual presentation of PAN diagnosed with multidisciplinary input from the urology, radiology and nephrology teams.

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Polyarteritis nodosa (PAN) is an uncommon systemic vasculitis characterized by necrotizing inflammation of small- or medium-sized arteries. It was the first vasculitis to be described by Kussmaul and Maier in 1866 and was named "periarteritis nodosa."[1]

Signs and symptoms of this disease are usually vague and non-specific. Malaise, weakness, fever, headache, arthralgia, myalgia, chronic renal failure, neuropathy and transient ischemic attacks are common presentations. Angina, myocardial infarction and congestive cardiac failure are rarely seen at the first presentation. Abdominal pain with nausea, vomiting or per rectal bleeding, as well as hepato-pancreatic infarction, have also been described. Many clinical symptoms are related to arterial branch occlusion leading to organ ischemia.[2] The lungs are usually not involved in systemic PAN.[3] It affects 2 to 6 people per 100 000 per year, and can be seen in all ethnic groups. Any age group can be affected, but it is commonly seen in people between the ages of 40 and 60. The incidence is higher in areas where hepatitis B is endemic.[4]

Common investigations to diagnose PAN include positive anti-neutrophil cytoplasmic antibodies (ANCA), hepatitis B surface antigen and a raised erythrocyte sedimentation rate (ESR). A full blood count shows raised inflammatory parameters (neutrophils) and gamma globulins.[5] Angiography shows micro aneurysms, which is considered the gold standard for diagnosis.[6]

We report the case of a 65-year-old man who was diagnosed to have polyarteritis nodosa having presented atypically with hematuria. He also had a testicular lesion which was initially presumed to be a tumourous growth.

The patient had a history of myelodysplasia (chronic myeloid leukemia), which is currently in remission and under regular follow-up. After being referred by hematologists with a suspected left testicular lesion, he visited the urology clinic. The lesion was cystic on examination and initial imaging with ultrasound suggested the possibility of mycotic aneurysmal lesions in the testicle (Fig. 1).

While he was being worked up for the suspected testicular lesion, he developed frank painless hematuria, which was initially managed with catheterization and bladder irrigation. His condition then deteriorated and he became acutely septic with respiratory compromise. Acute severe pneumonia was diagnosed which required prolonged ventilation. During the course of his intensive care unit admission, in spite of anti-

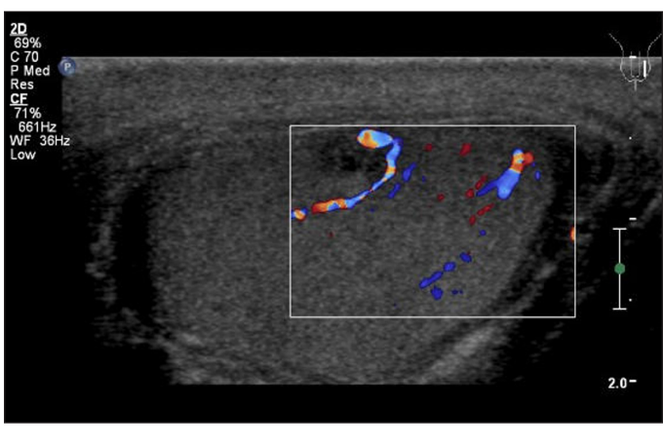

Fig. 1. A testicular ultrasound, suggesting the possibility of mycotic aneurismal lesions.

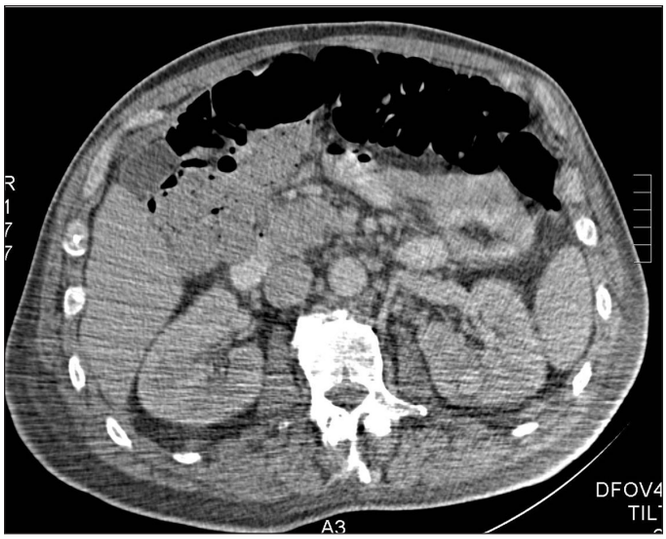

Fig. 2. A computed tomography scan showing a renal parenchymal aneurysm.

biotic administration, his inflammatory parameters remained high. Hematuria continued requiring intermittent bladder irrigation and supplemental blood transfusions.

Once his general condition improved, he had a cystoscopic evaluation. There were no gross abnormal lesions in the bladder, but blood was observed coming out of the left ureteric orifice. Ultrasonography suggested a left renal hematoma confirmed on a subsequent computed tomography scan (Fig. 2), which also showed small aneurysms in both kidneys. Renal angiography was undertaken with a view to embolization of any bleeding lesion. Angiography revealed numerous aneurysms in both kidneys measuring up to 15 mm in diameter (Fig. 3). Segmental infarcts were

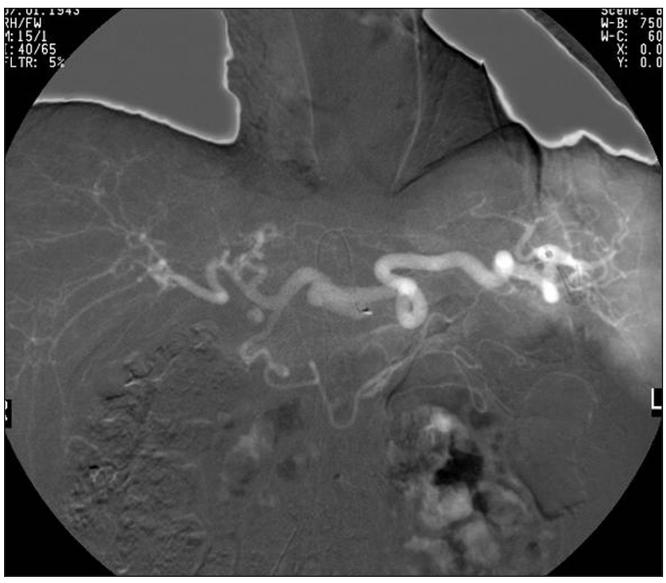

Fig. 4. The mesenteric angiogram showing an aneurysm in the pancreatic head.

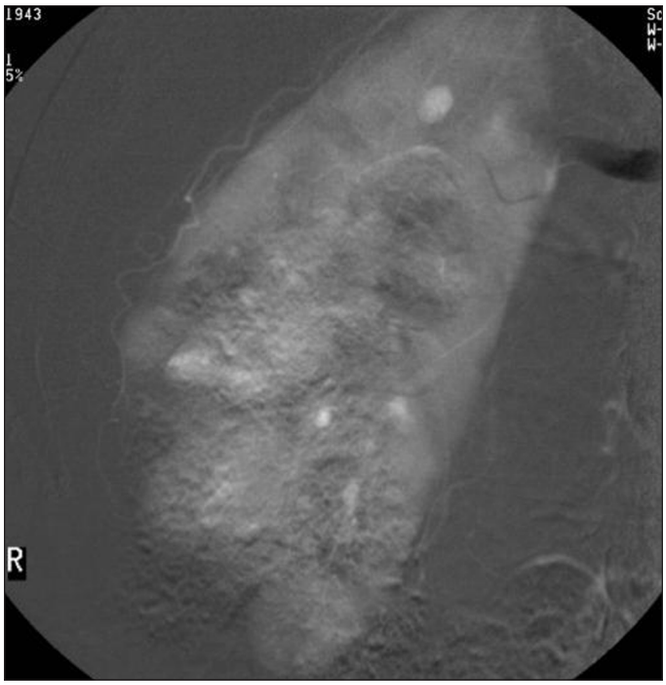

Fig. 3. The renal angiogram revealing multiple aneurysms and segmental infarct.

also noted in the lower pole of the right kidney. Further mesenteric angiography revealed small aneurysms in the head of the pancreas (Fig 4). Subsequently laboratory studies showed positive ANCA and raised ESR levels. Taken together, these findings suggested the diagnosis of PAN, and the patient was started on corticosteroid therapy. He made a good clinical recovery.

Currently, he is on a reducing dose of steroid treatment and remains clinically well. His management continues under the nephrology and hematology teams for his PAN and myelodysplasia, respectively.

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